

## 5.6 (old 3.13.6) Optional Hemoglobin Follow-up – FCD, FCE, FCV, FDC, FDE, FDV, FEC, FED, FEV, FVD, FVE, and Patterns with 4-5 Hemoglobins

### GENERAL INFORMATION:

- This category includes hemoglobin patterns with:
  - combinations such as FCD which occur infrequently and for which there is little or no information about the hemoglobin type being associated with significant clinical consequences
  - more than three significant hemoglobin peaks on the newborn screening hemoglobin chromatogram. Results with 4 to 5 hemoglobins are considered uninterpretable. Most of these patterns contain Hb A and after further testing are found to be unidentified hemoglobin variant traits.
- Follow-up for these results is optional. The primary care provider (PCP) can contact the ASC for confirmatory testing and consultation with a hematologist.

- POLICY:**
1. If the PCP chooses to pursue follow-up for infants with the above patterns, he/she may contact the ASC NBS Coordinator and this protocol will be followed.
  2. E-mails drafts of doctor letters to NBSB Hb Coordinator for review, to assure consistency state-wide.

### PROTOCOL:

Resp. Person	Action
Results mailer, indicating unusual hb pattern	<ul style="list-style-type: none"><li>• No interpretation possible due to unusual pattern; further testing available by request.</li><li>• Instructs physician of record to contact ASC listed on the results mailer for assistance/consultation.</li></ul>
ASC NBS Coord.	<ul style="list-style-type: none"><li>• If confirmatory testing is requested by the PCP, assists in arranging for blood specimens to be drawn from the infant and both parents, if possible, at the birth hospital lab or other collection site and shipment to the Hemoglobin Reference Lab at Children's Hospital &amp; Research Center at Oakland (CHRCO).</li></ul>

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	<ul style="list-style-type: none"> <li>• Opens case in SIS in order to document contacts with the PCP and follow-up on confirmatory testing.</li> <li>• Sends <i>Instructions for Collection, Handling, And Mailing Of Blood Specimens For Confirmatory Testing</i> (5.8), as well as the shipping materials (cylinder, GSO label) to lab obtaining confirmatory specimen(s).</li> </ul>
Hemoglobin Reference Lab (CHRCO)	<ul style="list-style-type: none"> <li>• Conducts confirmatory testing on liquid blood specimen(s).</li> <li>• <b>Within 11 working days</b> enters results in SIS and informs the ASC NBS Coordinator of the confirmatory test results by fax or phone, followed by a hard copy sent to the ASC NBS Coordinator and NBSB Hemoglobin Coordinator. Includes the following results: <ul style="list-style-type: none"> <li>A) Separation of hemoglobins F, A, S, C, D, and E with relative concentrations for each hemoglobin on all specimens by cellulose acetate-citrate agar electrophoresis, isoelectric focusing, high pressure liquid chromatography, and/or DNA analysis as outlined in the Hb Reference Lab NBS vendor agreement scope of work or as approved by the NBSB.</li> <li>B) Hemogram on each suitable specimen, to include hemoglobin, hematocrit, and mean corpuscular volume (MCV) and mean corpuscular hemoglobin (MCH).</li> <li>C) Free Erythrocyte Protoporphyrin (FEP) on specimens with microcytic hypochromic anemia.</li> <li>D) Quantitative Hb A2 when necessary to resolve phenotype.</li> <li>E) Quantitative Hb F when necessary to resolve phenotype.</li> </ul> </li> <li>• Informs the ASC NBS Coordinator of the test results by fax and sends a hard copy to the ASC NBS Coordinator and NBSB Hb Coordinator.</li> <li>• <b>Within 31 calendar days of receipt of specimen(s)</b>, faxes and mails reports on results of DNA analysis, if needed to determine the hemoglobin type (only if no Hb A is present in the infant or both parents carry a hemoglobin variant trait).</li> </ul>
NBSB Hb Coord.	<ul style="list-style-type: none"> <li>• Enters confirmatory Hb results in SIS.</li> </ul>
ASC NBS Coord.	<ul style="list-style-type: none"> <li>• <b>Phones the NBSB Hb Coordinator with any confirmatory results that are inconsistent with the NBS results prior to contacting the PCP.</b></li> <li>• Phones the PCP with the confirmatory test results.</li> <li>• Provides contact information for the ASC's consulting hematologist (in addition to the Hemoglobin Reference Lab consulting hematologist listed on</li> </ul>

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ASC NBS Coord.	<p>the lab report) to the physician for discussion of the test results and recommendations, if requested.</p> <ul style="list-style-type: none"><li>• Consults with the NBSB Hemoglobin Coordinator about wording for the confirmatory hemoglobin results letter to the physician.</li><li>• If the baby is evaluated at a CCS Sickle Cell Disease Center, refers the case to the Center in SIS. If the Center has an NBS Vendor Agreement, the Center should enter a Hemoglobin Service report with the “test resolution” and “Disorder” for the ASC to resolve the case.</li><li>• For results of unknown hemoglobin variant trait or an uncommon hemoglobin trait, parents can be referred for genetic counseling at a Sickle Cell Disease Center or Prenatal Diagnosis Center.</li><li>• Enters case notes in SIS. If the baby was not seen at an NBS- vendored CCS Sickle Cell Disease Center, resolves case on Case Resolution page in SIS as “Other Conditions” or “Carrier” depending on confirmatory results interpretation. Selects appropriate Follow-up Status and notes interpretation of confirmatory results in Comment box. (Or resolves as lost to follow-up, parent refusal, newborn expired, or no response from parents, if applicable).</li></ul>
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